Successful treatment tailored to each splanchnic arterial lesion due to segmental arterial mediolysis (SAM): Report of a case

Takuya Hashimoto, MD,[a] Juno Deguchi, MD,[a] Hisako Endo, MD,[b] and **Tetsuro Miyata, MD,[a]**
Tokyo, Japan

Segmental arterial mediolysis (SAM) is a rare condition characterized by splanchnic arterial catastrophe caused by mediolysis. We report a 59-year-old man with a ruptured splenic arterial aneurysm who was successfully treated by coil embolization. He underwent additional resection of large gastroepiploic and residual splenic aneurysms. Pathological examination showed mediolysis and tearing, compatible with SAM. Furthermore, he developed acute dissection of the superior mesenteric artery (SMA) one and a half years later, demonstrated by computed tomography. This report demonstrates that SAM is characterized by multiple lesions of the splanchnic arteries at different times, and requires treatment suited to the lesions, including careful long-term observation. (J Vasc Surg 2008;48:1338-41.)

Segmental arterial mediolysis (SAM), first described by Slavin et al[1] in 1976, is a non-atherosclerotic, non-vasculitic disease with wide variation in the clinical course, requiring pathological evaluation. The disease usually affects middle-sized splanchnic arteries, and is characterized by lysed smooth muscle cells within the arterial media with surrounding fibrosis.[2-7] It can cause dissection, aneurysm, or stenosis of multiple splanchnic arteries, resulting in sudden abdominal pain or distention, which frequently needs emergency surgical intervention. However, the clinical course of this disease, including the development of new lesions, remains elusive, although patients with SAM often develop lesions of multiple splanchnic arteries simultaneously.

SAM is characterized by mediolysis and tearing separation from the adventitia, as well as surrounding fibrosis of the splanchnic arteries. It is still controversial whether SAM is a distinct disease entity or a variant of fibromuscular dysplasia (FMD) in which fibrosis is prominent.

We report a patient with SAM who underwent staged surgery for multiple aneurysms. Moreover, he developed dissection of the SMA during follow-up, which was treated conservatively and resulted in thrombosis of the false lumen.

CASE REPORT

A 59-year-old man with sudden onset of abdominal pain was transferred to our hospital in a state of shock. He had been healthy without any cardiovascular risk factors including hypertension, smoking, diabetes, and hyperlipidemia.

On initial physical examination, he was conscious with systolic blood pressure of 64 mm Hg. Heart rate was elevated to 120 beats per minute. Most laboratory data on admission were unremarkable; including white blood cell count of $3.9 \times 10^3/mm^3$, red blood cell count of $530 \times 10^4/mm^3$, and platelet of $251 \times 10^3/mm^3$. Coagulation panel was within normal limits. Electrocardiography and cardiac enzyme examination ruled out myocardial infarction.

Computed tomography (CT) showed multiple splenic artery aneurysms and a hematoma, measuring about $14 \times 10 \times 9$ cm, which developed around the largest aneurysm and extended into the left retroperitoneal space (Fig 1, *A*). It also depicted bilateral aneurysms of the renal arteries. Emergency embolization of the splenic artery with microcoils achieved complete arrest of extravasation. He was transfused 20 units of blood and recovered uneventfully.

Because a follow-up CT scan three weeks later detected aneurysms of the gastroepiploic artery, the patient underwent complete selective splanchnic angiography, which newly indicated multiple aneurysms involving the gastroepiploic and the distal splenic artery at the hiatus of the spleen (Fig 1, *B*), as well as the bilateral renal aneurysms which remained unchanged in size (Fig 2, *A, B*). Aneurysms of the gastroepiploic artery, up to 2 cm in size, showed a beaded appearance and were interspersed with focal segmental stenosis. Collaterals from the superior mesenteric artery (SMA) persistently supplied arterial flow to the distal splenic arterial aneurysms. Moreover, a head CT scan revealed a small saccular aneurysm of the right internal carotid artery (Fig 2, *C*). An echocardiogram showed normal movement of cardiac wall with 65% of the ejection fraction rate.

He underwent resection of the gastroepiploic artery together with exclusion of the distal splenic artery to prevent rupture of the aneurysms. He recovered uneventfully and was discharged from our hospital.

Histological examination of the resected arterial wall specimen detected so-called 'medial islands' in extensive fibrous granulation in the aneurysmal wall (Fig 2, *D*). There was fibrous granulation in

From the Division of Vascular Surgery, Department of Surgery,[a] and the Department of Pathology,[b] Graduate School of Medicine, The University of Tokyo.

Competition of interest: none.

Correspondence: Tetsuro Miyata, MD, Division of Vascular Surgery, Department of Surgery, Graduate School of Medicine, The University of Tokyo, 7-3-1 Hongo, Bunkyo-ku Tokyo, 113-8655 Japan (e-mail: miyata-2su@h.u-tokyo.ac.jp).

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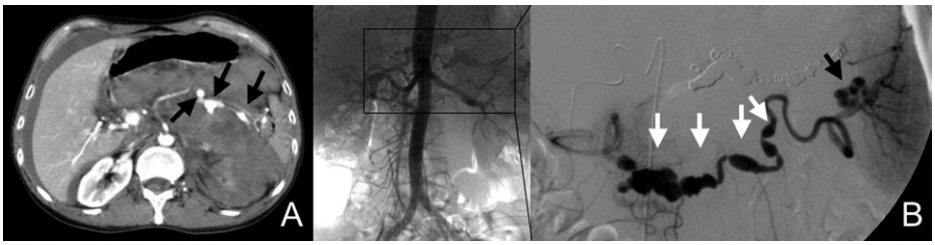

Fig 1. A, CT of the abdomen shows a large fluid collection in the left anterior pararenal space. Note the beaded aneurysms of the splenic artery (*arrow*). **B,** Selective angiography of the gastroepiploic artery. Saccular aneurysms and multiple stenotic regions are seen. There are also residual splenic artery aneurysms (*arrow*) at the distal site of coil embolization (*white arrow*).

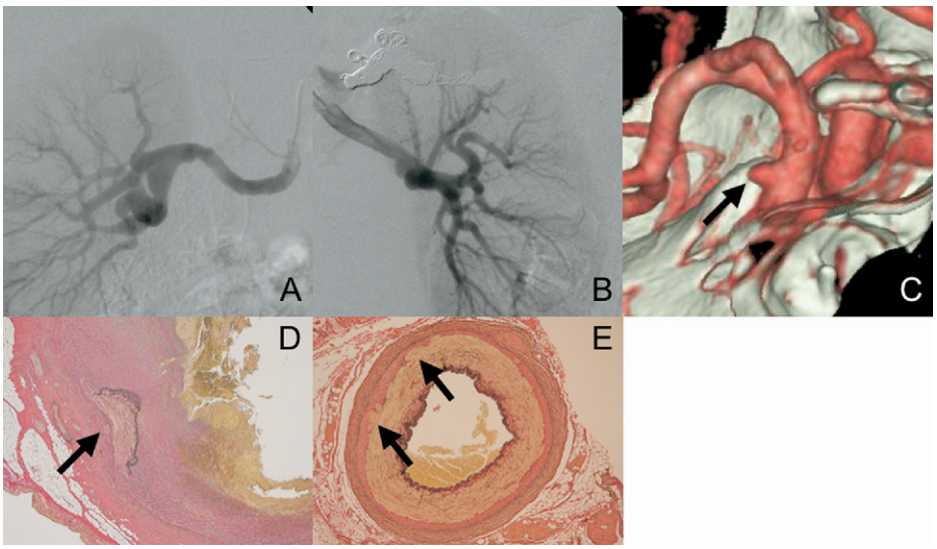

Fig 2. A, B, Angiography of renal arteries. The bilateral renal arteries show aneurysmal change. **C,** 3-D CT of the head and neck. A small aneurysm (3 mm in diameter) is demonstrated in the distal site of the right internal carotid artery (*arrow*). **D,** Aneurysmal lesion of gastroepiploic artery. *Arrow* indicates a medial island spared from mediolysis. Other muscle fibers are lysed and replaced by fibrous granulation tissue. The internal elastic lamina is almost lost except for a medial island (Elastica van Gieson stain; magnification ×20). **E,** Vessel wall without aneurysmal change. Segmental areas of the outer media show 'lytic spaces' in which smooth muscle cells appear to have been lysed, showing an irregular border and replacement by fibrous granulation tissue (*arrow*) (Elastica van Gieson stain; magnification ×10).

the outer media, with separation from the adventitia in artery without aneurysmal change (Fig 2, *E*). The medial layer showed no evidence of inflammation. The adventitia contained a few leukocytes. There was no evidence of atherosclerotic change, mycotic aneurysm, or vasculitis.

One and a half years later, the patient presented to our hospital with upper abdominal pain with acute onset. A CT scan depicted a dissection of the SMA without decreased circulation to the intestine through the true lumen, while the false lumen was almost completely thrombosed. He recovered without any intervention. The bilateral renal artery aneurysms remained unchanged. Echography showed that the false lumen of the dissected SMA was completely thrombosed 11 months later. Fig 3 summarizes the time course of the results of all angiographic studies. These vascular lesions have been under close observation. Long-term and careful observation is necessary for SAM, because of its progression as well as the development of new lesions, the so-called 'spatiotemporal' property of SAM.

DISCUSSION

Slavin first described three autopsy cases of segmental mediolysis in the abdominal muscular arteries in 1976 and advocated a pathologically distinct disease entity, 'segmental arterial mediolysis; SAM'. SAM is characterized by involvement of multiple splanchnic arteries with no involvement of extra-abdominal arteries. The mediolysis of arteries results in aneurysm, dissection, or stenosis, causing rupture or acute interruption of the arteries, necessitating emergency surgical intervention.

The etiology of SAM is still unclear. Slavin et al have postulated that repeated vasoconstrictive responses to

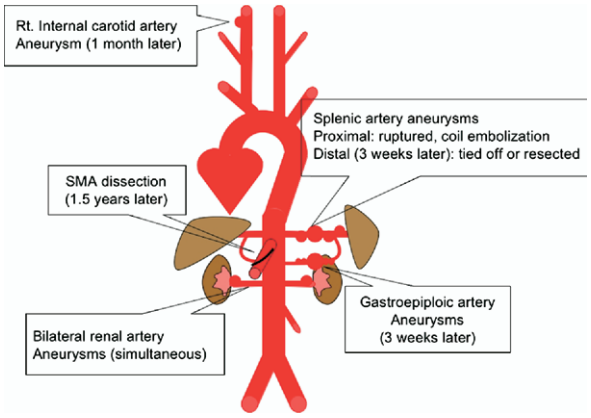

Fig 3. Time course of vascular events. Aneurysms of the bilateral renal arteries were already noted at the time of rupture of splenic artery aneurysms. Gastroepiploic artery aneurysms and carotid artery aneurysm were detected 3 weeks and 1 month later, respectively. One and a half years elapsed from the initial event to the onset of SMA dissection.

shock or severe hypoxemia in the splanchnic vascular bed may result in arterial mediolysis, because arteries with chronic vasospasm have histological similarities to segmental arterial mediolysis.[3,4] The present patient, however, had no history prior to the onset of SAM of co-existing diseases that are associated with vasoconstrictor stimuli such as Raynaud's disease, migraine, hypertension, pulmonary hypertension, shock, stroke, and recent surgery.

The differential diagnoses of mycotic aneurysm and arteritis should be considered in patients with multiple splanchnic aneurysms. It was, however, improbable that our patient had a mycotic aneurysm or vasculitis because there was no evidence of inflammation on both physical examination and laboratory data. We also ruled out Takayasu's arteritis which usually affects more proximal part of splanchnic vessels.

The present patient underwent emergency coil embolization for the ruptured splenic artery, followed by screening of the splanchnic artery system. Subsequently, he underwent surgical resection of the gastroepiploic artery aneurysm. It is also noteworthy that our patient developed dissection at the SMA one and a half years after rupture of the splenic artery. The angiographic findings of SAM are irregularity of the vascular wall, widening and narrowing of a vessel (string of beads appearance), an occlusion or interruption, and dissection or aneurysmal enlargement.[2,8-10] In addition, a hematoma around affected vessels is often shown on CT (much more easily visible on MDCT).[11] However, those findings seen in SAM are often similar to those of vasculitis such as polyarteritis nodosa, fibromuscular dysplasia, and pseudoaneurysms, making its differential diagnosis much more difficult. It is known that these angiographic findings are a reflection of the histological changes. It is also reported that these findings show changes during follow-up (dilatation, beading with narrowing and restoration of the smooth wall, with various modifications such as aneurysmal enlargement and occlusion).[9] Hemodynamic alterations may also affect these changes. In our case, the gastroepiploic artery, which was initially normal, developed a string of beads appearance and aneurysmal dilatation. There are some reported cases of imaging follow-up of pre-existing lesions of SAM, which show various degrees of remodeling of the arterial lumen with diminished or increased size of aneurysms.[9,10,12] However, no report has mentioned the development of new lesions in the follow up of SAM. SAM can affect multiple muscular arteries spatiotemporally, and close attention to new lesions is needed in the course of follow-up.

In conclusion, in cases of abdominal artery aneurysms, physicians should keep the possibility of SAM in mind, and screening of the abdominal and extra-abdominal arteries is recommended after emergency treatment. When residual aneurysms are detected, treatment should be tailored according to the severity of each aneurysm. Long-term careful observation is mandatory for SAM, because of its progression as well as the development of new lesions, its so-called 'spatiotemporal' character.

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AUTHOR CONTRIBUTIONS

Conception and design: JD, TM
Analysis and interpretation: YI, HE
Data collection: TH, YI, HE
Writing the article: TH
Critical revision of the article: JD, TM
Final approval of the article: HN
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